中文題目:頑強的便秘—成人型大腸神經節減少症

英文題目：Refractory constipation in a healthy man – Hypoganglionosis in adults

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Background：The Hypoganglinosis is a rare and resembles Hirschsprung's disease in the male-to-female ratio, etiology and clinical presentation. Although chronic constipation is a common complaint in general population, the patients with constipation can accompany with fatal complication if the patient with megacolon which is related with hypoganglinosis. Up to 90.5% of patients with Hirschsprung's disease are diagnosed in the newborn period and the median age at diagnosis of the patient with hypoganglinosis was 4.85 years old. We report a healthy 43 year-old male with severe constipation who is diagnosed as adult type hypoganglinosis.

Methods：A 43-year-old man presented with chronic constipation, abdominal discomfort, and flatulence, and had been passing intestinal gas with unusual odors for 18 months.

Computed tomography showed a severely dilated feces-filled proximal segment of the descending colon and transverse colon. The patient underwent subtotal colectomy with ileocolic anastomosis for a clinical impression of intractable megacolon. Histopathology confirmed a diagnosis of hypoganglionosis of the colon.

Discussion：Chronic constipation is a common complaint in the general population; however, when segmental or total colonic distension of > 6 cm is seen on a plain film, toxic megacolon should be considered because it is a potentially fatal complication. The differential diagnosis of megacolon includes mechanical obstruction,

postoperative ileus, paralytic ileus, ulcerative colitis, Crohn's disease, infection, ischemia, malignancy, hypokalemia, hypomagnesemia, hypothyroidism, colonic distension (intestinal pseudo-obstruction), and any drugs that slow colonic motility.(1, 2). Aside from toxic megacolon, functional constipation with bowel distention can be divided into 2 groups based on the site of dilatation – megarectum or megacolon. Mega-bowel might occur secondary to intestinal dysganglionoses, anorectal obstruction, or disorders of the endocrine and central nervous systems. The gold standard of diagnosis remains a full-thickness biopsy of the bowel.

The intestinal dysganglionoses is one of adult intestinal innervations disorders, represent a heterogeneous group of enteric nervous system anomalies, such as adult Hirschsprung's disease, intestinal neuronal dysplasia, internal anal sphincter neurogenic achalasia and hypoganglionosis.(3, 4)

Diagnostic tests for adult hypoganglionosis include contrast enema, anorectal manometry, and rectal suction biopsy with hematoxylin and eosin and acetylcholinesterase stain. There is no significant difference between Hirschsprung's disease and hypoganglionosis as observed on barium enema.(5) Both present with a narrow distal segment and dilated proximal segment, and some are found to have classic funnel-shaped dilation at the level of the transition zone.

Conclusion：The hypoganglinosis is rare disease in population, especially in adult group. However, the early and accurate diagnosis of adult hypoganglionosis is important for the physician to avoid fatal complication. We hope that adult intestinal innervation disorders will be considered readily in the differential diagnoses of chronic constipation.

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